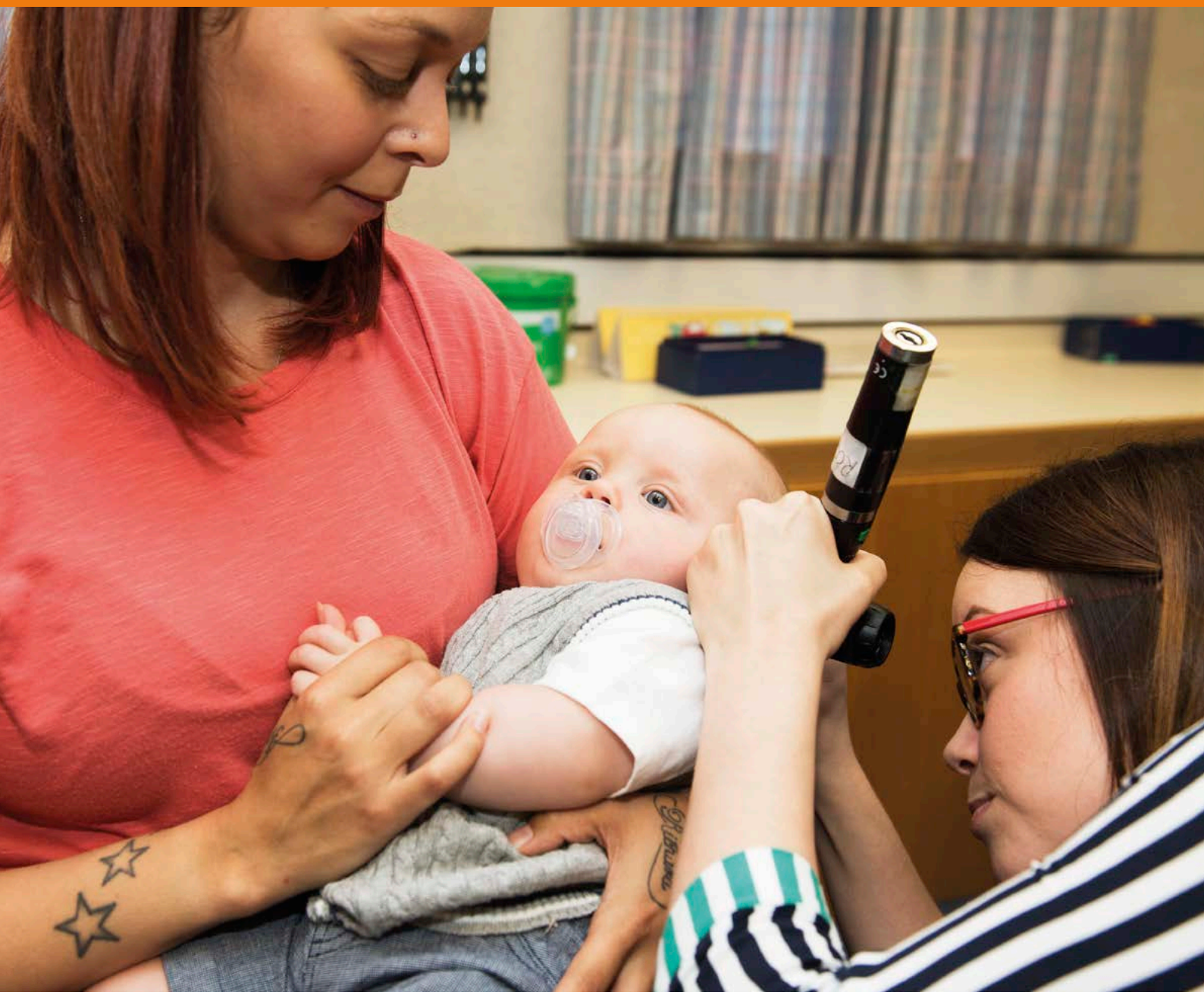
Understanding your child's hearing tests

Information for families

A guide to the hearing and medical tests that are used to find out the type, level and cause of deafness



ndcs
every deaf child

Our vision is a world without barriers for every deaf child.

Acknowledgements

We would like to thank the members of the Aetiological Investigations Group for the valuable advice they gave to help us produce the information in chapter 8.

Thanks to Contact a Family for letting us reproduce the information from their resource *Finding Medical Information on the Internet* in chapter 9.
www.cafamily.org.uk/medical-information

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We use the term 'deaf' to refer to all types of hearing loss from mild to profound. This includes deafness in one ear or temporary hearing loss such as glue ear.

We use the term 'parent' to refer to all parents and carers of children.

Throughout this resource we use the following terms:

- 'your doctor' to mean the doctor in charge of your child's audiological care. This may be an audiovestibular physician, ear, nose and throat (ENT) specialist, or community paediatrician in audiology
- 'your family' includes your child's grandparents on both sides.

1. Introduction

This resource explains the different types and levels of deafness and has information on the different tests that can check a child's hearing. It explains about the ear and how it works, different types of deafness, and audiograms (a chart on which your child's hearing test results will be written). It also has information on the different medical tests or investigations that are used to help diagnose the cause of permanent deafness.

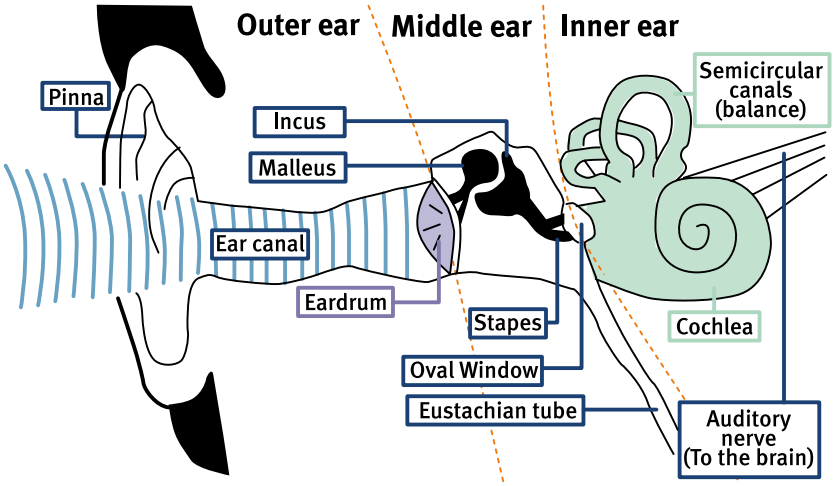
Throughout this resource we'll direct you to some of our other information resources which are available to download from our website at **www.ndcs.org.uk.** For more information and practical support or to order copies of other resources, phone our Freephone Helpline on 0808 800 8880, email us at **helpline@ndcs.org.uk** or contact us via live chat at **www.ndcs.org.uk/livechat.**

2. The ear and how it works

The ear has two main functions.

- It receives sound and changes it into signals that the brain can understand.
- It helps us to balance.

The two functions are closely related.



Ear diagram

The ear

The ear is the first part of the hearing system. The pinna (the outside part of the ear) collects sound waves, directing them down the ear canal. The waves then cause the eardrum to vibrate. These vibrations are passed across the middle ear by three tiny bones: the malleus, incus and stapes (sometimes known as the hammer, anvil and stirrup, known together as the ossicles). The bones increase the strength of the vibrations before they pass through the oval window into the cochlea.

The cochlea looks like a snail's shell. It's filled with fluid and contains thousands of tiny sound-sensitive hair cells. The vibrations entering the cochlea cause the fluid and hair cells to move, much like the movement of seaweed on the seabed when waves pass over it.

As the hair cells move, they create a small electrical charge or signal. The auditory nerve carries these signals to the brain where they are understood as sound.

For an ear to work fully and allow us to pick up sound, all of these parts must work well. Deafness happens when one or more parts of the system are not working effectively.

Balance (vestibular) testing

Balance

The brain uses information from the eyes (what we see), our body (what we feel), and the inner ear to balance. The semicircular canals in the inner ear are three tubes, filled with liquid and movement-sensitive hair cells. As we move, the fluid moves. This creates signals that are sent to the brain about balance.

For more information about balance visit **www.ndcs.org.uk/balance**

3. Different types of deafness

There are three types of deafness: conductive, sensorineural and mixed.

Conductive deafness is when sound can't pass efficiently through the outer and middle ear to the cochlea and auditory nerve. There are several possible causes, including impacted wax, an ear infection and underdevelopment of the outer ear, ear canal or middle ear.

The most common type of conductive deafness in children is caused by glue ear, with 8 out of 10 children having a bout of it before the age of 10 years. It's most common in young children, affecting about 1 in 5 pre-school children at any time. Glue ear (also known as OME – otitis media with effusion) is a build-up of fluid in the middle ear. For most children, the glue ear clears up by itself and doesn't need any treatment. For some children with long-term or severe glue ear, hearing aids may be provided, or the child may need surgery to insert grommets into the eardrums. Grommets are tiny plastic tubes which allow air to circulate in the middle ear. This keeps the pressure on either side of the eardrum equal which helps to stop fluid from building up.

For more information visit **www.ndcs.org.uk/glueear**

Sensorineural (or nerve) deafness is when there's a fault in the inner ear (most often because the hair cells in the cochlea are not working properly) or auditory (hearing) nerve. Sensorineural deafness is permanent.

Mixed deafness is when there's a combination of sensorineural and conductive deafness, such as when a child has glue ear and a permanent sensorineural deafness.

4. Hearing tests

There are different tests that can be used to find out how much hearing your child has. The tests used will depend on your child's age and stage of development. It's possible to test the hearing of all children from birth. Screening tests are normally done first and if it's likely that there's a hearing loss, the child will be referred to an audiologist. The audiologist will then perform a range of objective and/or behavioural tests to build up an accurate picture of your child's hearing.

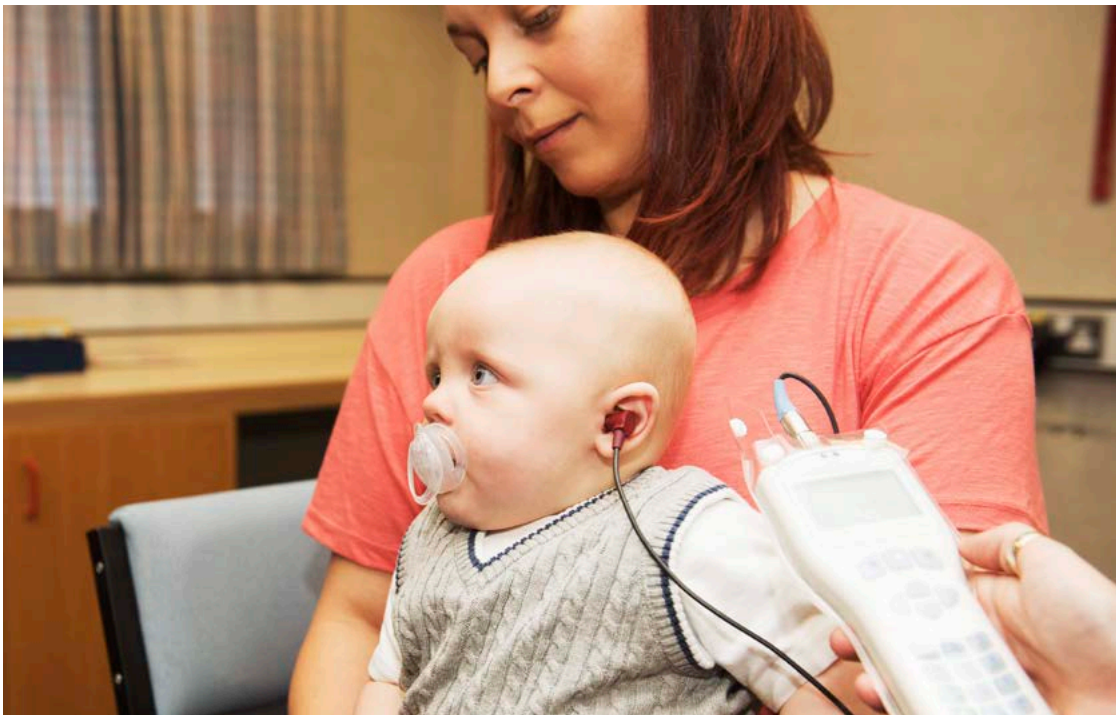
Objective hearing assessments use test techniques that measure the function of the ear or hearing nerve pathways. They don't require a response from the child to show that they have heard the sound.

Behavioural hearing assessments use test techniques where the audiologist records the child's response to sound. This might be a startle, look or head turn in a baby, or an older child moving a toy or saying "yes" when they hear a sound.

You can ask your audiologist for a copy of your child's test results. You may like to keep them at home or take them with you when you visit the audiology department or ear, nose and throat (ENT) doctor.

Speech discrimination testing – the McCormick Toy Test

Objective hearing tests



Otoacoustic emissions (OAE) testing

Otoacoustic emissions (OAE)

The OAE test works on the principle that a healthy cochlea (inner ear) will give a faint response when stimulated with sound. The cochlea contains outer hair cells which produce this response to sound. The OAE test gives information about how well the outer hair cells are working.

A small ear piece, containing a speaker and a microphone, is placed in the baby's ear. A clicking sound is played and if the cochlea is working properly the microphone in the ear piece will pick up the response. This is recorded on a computer that tells the screener if the baby needs to be referred for a further screening test. If the test records strong responses from the baby's ear then they will not need any more tests.

Although it's a test of how the ear is working, the OAE test can't give information about levels of hearing loss. The screening test is very quick and gives the result straightaway.

A poor response to an OAE test doesn't necessarily mean that a child is deaf. It can be difficult to get a response if the baby is unsettled at the time of the test, if the room is noisy, or if there's any fluid in the ear from the birth. This test is usually repeated before referral for the second type of screening test, which is called auditory brainstem response (ABR).

A baby being prepared for auditory brainstem response (ABR) testing

Auditory brainstem response (ABR)

If deafness is suspected or your baby has failed the newborn hearing screening test, they will be referred to audiology for more tests.

The ABR measures whether sound is being sent from the cochlea and through the auditory nerve to the brain. It can be used as a screening test (automated auditory brainstem response – AABR – where the computer judges whether there's a response at quiet levels of sound). It can also be a more detailed test where different levels of sound are used and the audiologist interprets the results to find the quietest level of sound being picked up by the hearing nerves.

The audiologist will place three small sensors and a set of headphones on the child's head. For an accurate result, the child must be very still and quiet throughout the test. In young babies, the test is usually carried out while they are sleeping. In slightly older children, a light sedative or an anaesthetic may be offered.

In very young children or children who aren't developed enough to have behavioural hearing tests, the results of the ABR test can be used to accurately fit hearing aids if they are needed. In older children, this test may be used to confirm the results of their behavioural test.

Behavioural tests

As your child develops, their audiologist will assess their hearing using behavioural tests. These tests use toys and play as part of the assessment and involve your child listening for different sounds as part of a game.

Visual response audiometry (VRA) testing

Visual response audiometry (VRA)

Visual response audiometry is suitable for children from six months to about two-and a-half years. Using a machine called an audiometer, sounds of different frequencies and loudness are played through speakers. When the child hears the sound, they will turn their head and a visual 'reward' is activated, such as a toy lighting up or a puppet. The test can check the full range of hearing but doesn't give specific information about each ear. If your audiologist feels it's important to get information about each ear individually, this test can be done with small insert earphones.

Pure tone audiometry testing

Pure tone audiometry

From about the age of three, children are actively involved in testing by using a technique called conditioning. Younger children are shown how to move a toy (for example, putting a peg into a board) each time they hear a sound. Older children are asked to respond to sounds by saying yes or pressing a button. The sounds come through headphones, earphones placed inside the child's ear, or sometimes through a speaker (when the test is known as soundfield audiometry).

All of these tests are described as testing using air conduction – sounds passing through the ear canal and middle ear before reaching the cochlea. Auditory brainstem response, visual response audiometry and pure tone audiometry can also be tested using bone conduction.

Bone-conduction testing

Bone conduction

Bone-conduction testing is performed in a similar way to air-conduction testing. A small vibrating device is placed behind the child's ear. This allows sound vibrations to pass directly to the inner ear through the bones in the head, bypassing the outer and middle ear. This technique is useful for identifying whether a hearing loss is conductive or sensorineural.

Speech discrimination testing – the McCormick Toy Test

Speech discrimination tests

Speech discrimination tests check the child's ability to hear words at different listening levels. The tester asks the child to identify toys or pictures, or to copy words spoken by themselves or from a recording. From this the tester can assess the quietest level at which the child can correctly identify the words used. This test can also be used to assess lip-reading and signing skills.

An example of a speech discrimination test for younger children is the McCormick Toy Test. This test can be used for children aged 18 months and over. It's made up of 14 paired words which will be easily recognised by a child in the form of toys (see below list). The test can provide extra information to other hearing tests.

- tree – key
- shoe – spoon
- cow – house
- plane – plate
- horse – fork
- duck – cup
- man – lamb

The test starts with the tester asking the child to identify each toy. Any toy that isn't well known to the child is taken away (along with the other toy in the pair) and isn't included in the test. The test can be carried out with and without lip-reading and the words are spoken at conversational level. The child is taught to point to the toy when asked "show me the ...", "where is the...".

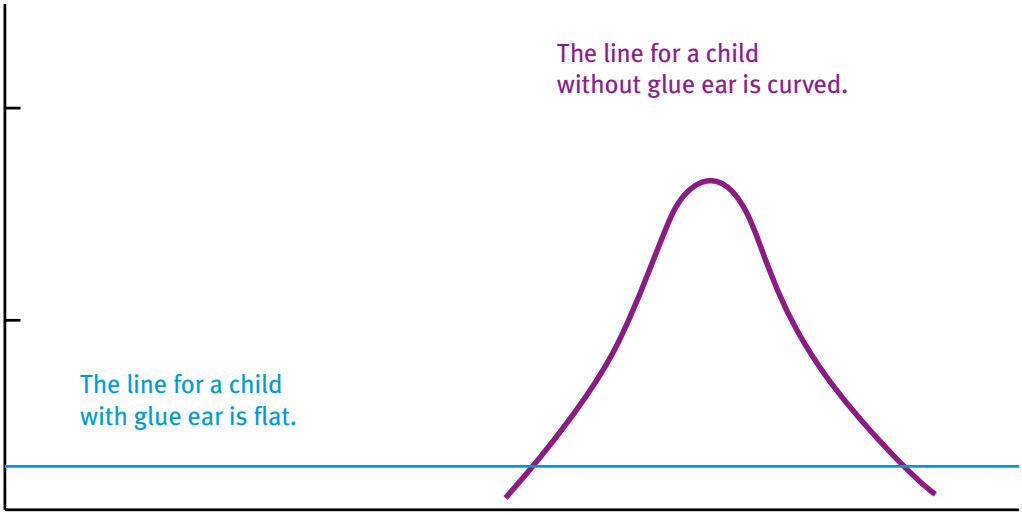
The tester then removes any visual clues by covering their nose/mouth with a small screen and repeats the test, lowering their voice until the child can correctly identify four out of five toys. The level of the tester's voice when the child got four out of five correct answers is then measured and recorded.

Tympanometry testing

Tympanometry

Tympanometry isn't a test of hearing; it's used to check how well the moving parts of the middle ear are working and your child will have this test as part of their hearing assessment.

A small earpiece is held gently in the ear canal. A pump causes the pressure of the air in the ear canal to change. The eardrum should move freely in and out with the change in pressure. The earpiece measures this by checking the sound reflected by the eardrum. If the eardrum isn't moving freely, there's likely to be some fluid or another problem with the middle ear. This build-up of fluid is usually glue ear (or OME – otitis media with effusion). Glue ear can cause temporary conductive deafness. For more information visit **www.ndcs.org.uk/glueear**



Tympanogram – a chart that shows how well the middle ear is functioning

Hearing tests and children with additional complex needs

The tests used will depend on your child's age as well as their stage of development. It should be possible to test the hearing of any child, whatever their stage of development, but it's more likely that several different tests will need to be done to get a clear picture of any hearing loss. Objective tests (such as OAE and ABR) don't need a child to respond to a sound in order to get a result. However, the child needs to be very still and quiet throughout the test, which may mean they need a light sedative or an anaesthetic.

Some children with additional needs may have to be tested using techniques that are normally used with younger children. If your local audiology service isn't confident about testing your child, they will refer you to another hospital or you can ask to be referred to another centre with more experience of testing children with complex needs.

Pure tone audiometry testing, using toys

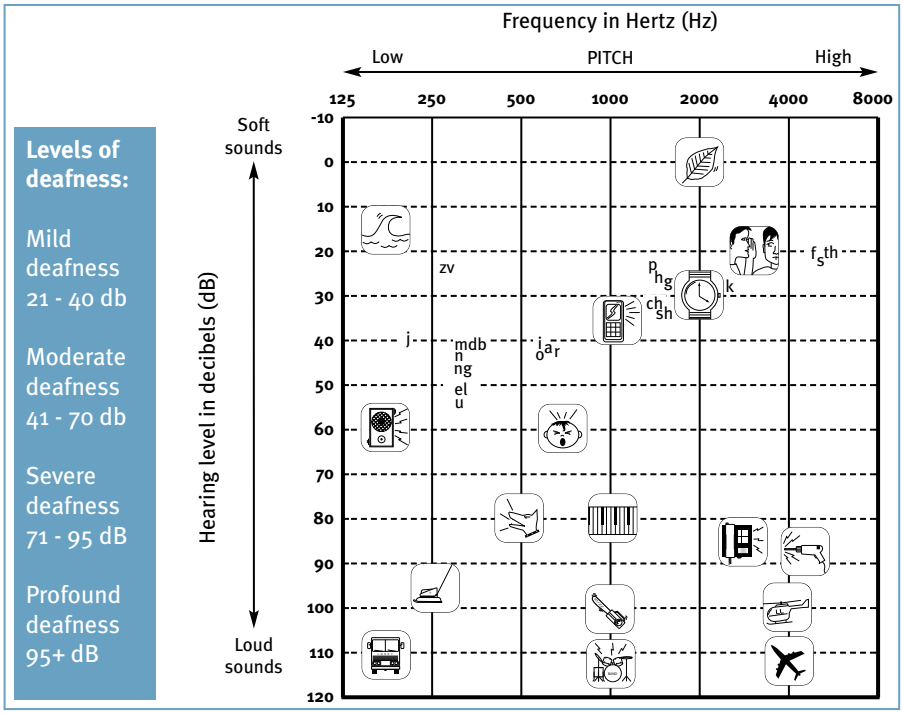
5. Audiograms

Some of your child's test results will be written on a chart like the one below, known as an audiogram. It shows you how loud a sound has to be, and at what frequency, before your child can hear it. Your child's test results may be plotted on one chart (as below) or two charts, side by side, for each ear separately. Crosses always show results for the left ear, and circles for the right ear. Your child may be deaf in one ear (unilateral deafness) or both ears (bilateral deafness). If your child is deaf in both ears, the deafness may be similar in both ears (symmetrical deafness) or different in each ear (asymmetrical deafness).

Your child's deafness may also be described as high frequency or low frequency, measured in hertz (Hz). We often think of frequency as the pitch of a sound. A piano keyboard runs from low-pitch sounds on the left to high-pitch sounds on the right and the audiogram is the same.

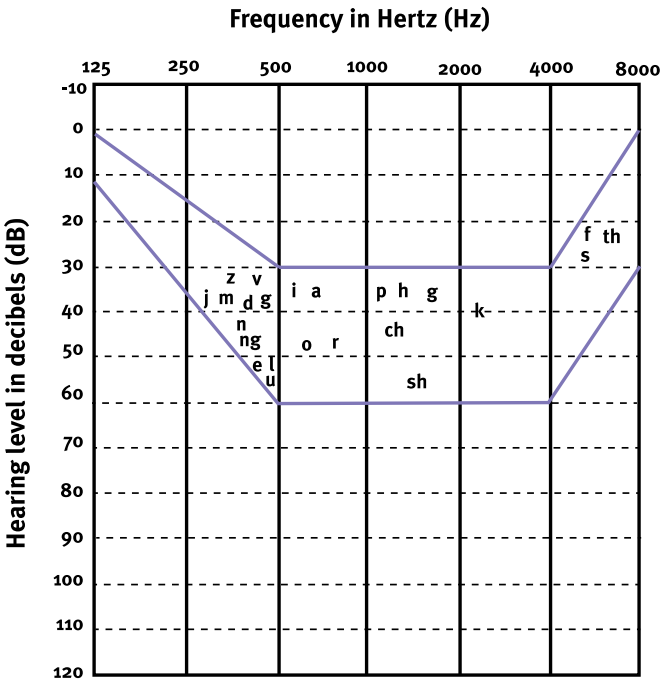
There are different levels of deafness. These can be described as a decibel hearing level (dBHL) (how loud a sound has to be for your child to hear it), or described using terms such as 'mild', 'moderate', 'severe' or 'profound'. The very quietest sounds are at the top of the chart, getting louder as you look down the page. With typical hearing, the lowest sounds people can hear are from 0–20dB.

Visual representation of the loudness and pitch of a range of everyday sounds



This diagram is based on British Society of Audiology definitions of hearing loss.

On the audiogram on the previous page, there are pictures of common sounds that give us an idea of loudness and frequency. There are also speech sounds from English drawn on the chart, and you can see that all the sounds of speech cover a range of frequencies. Try saying some of the speech sounds out loud while looking at the chart. The sounds m, b, and d are on the left-hand side and part way down the chart, meaning that they are lower frequency and slightly louder than say f, s, and th, which are higher in frequency and much quieter. The vowels also lie in the lower frequency range. Consonants tend to carry the meaning of the words and help with the intelligibility (understanding) of the sound. Try writing a short sentence using vowels only and then repeat the same sentence using consonants only. It's far easier to guess the words with the consonants than the words with only the vowels. Children with a hearing loss in the high frequencies mishear those vital consonants which carry the meaning of the word, making it more difficult to understand, particularly in background noise. So it's important to be able to hear sounds at a quiet level, across the frequency range, to be able to hear all the sounds of speech clearly.

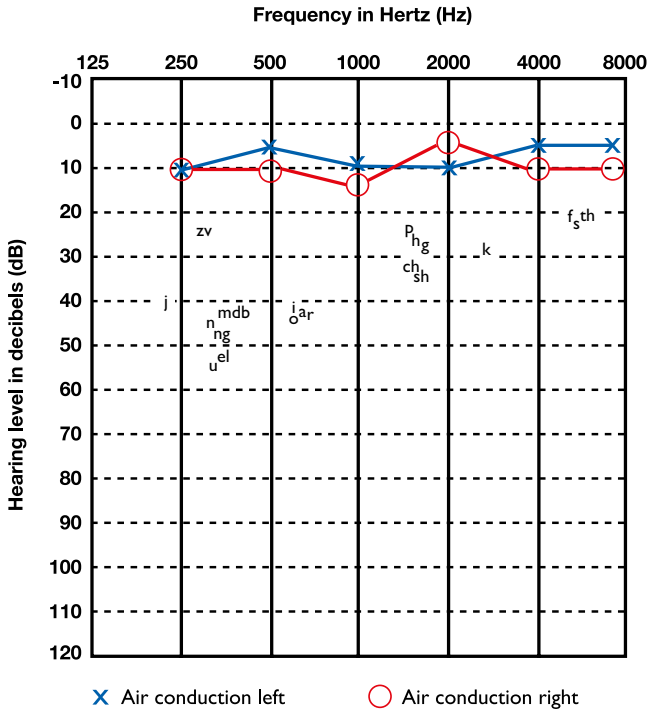


Audiogram showing a speech curve or 'speech banana'

When the speech sounds are plotted on an audiogram, they tend to fall within an area of frequencies forming a shape similar to that of a banana, hence the name 'speech banana' (also known as a speech curve). Most of the letters of the alphabet and letter combinations: 'ng', 'ch', 'sh' and 'th' lie within the speech banana. By looking at your child's audiogram, you can see which of the speech sounds your child can hear. Not being able to hear in those frequencies can affect a child's speech and language development.

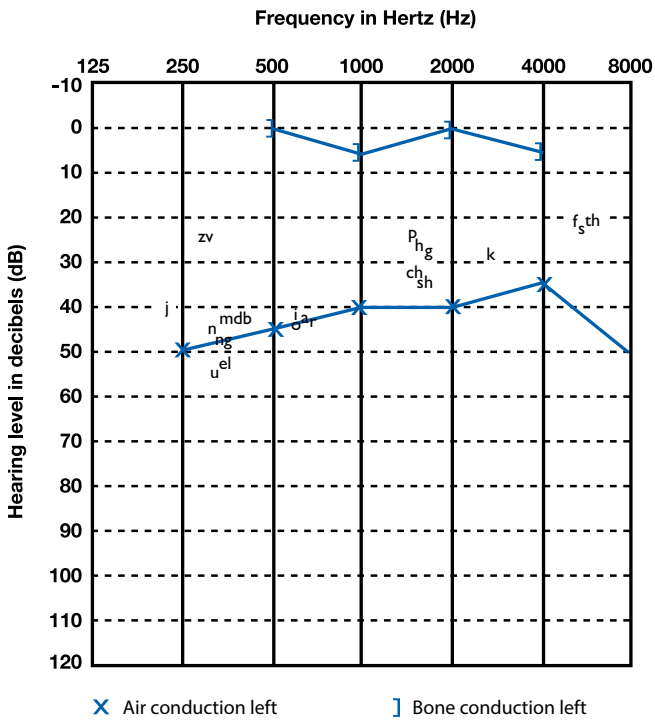
Ask your child's audiologist to explain your child's hearing test results to you and how they will affect your child's ability to hear speech.

Some examples of different audiogram results



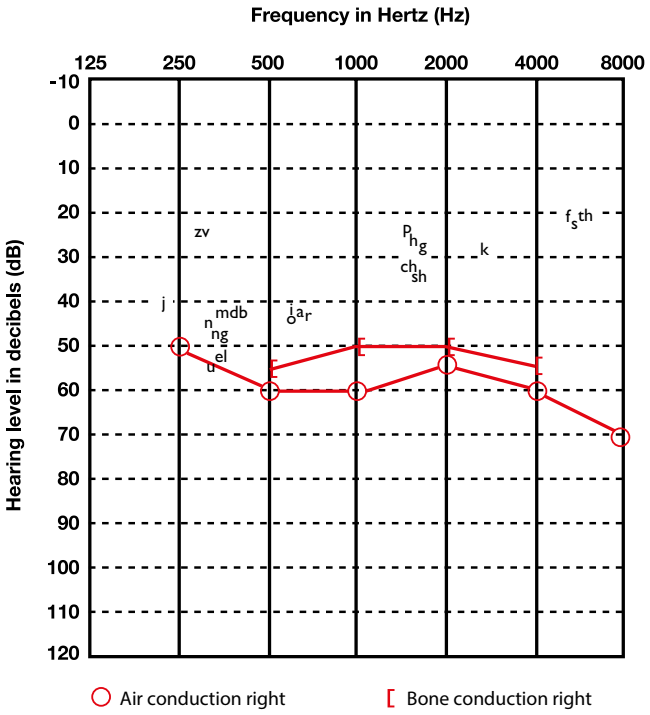
Typical range of hearing

This audiogram shows the level and range for a person with typical hearing levels.



Conductive deafness in the left ear

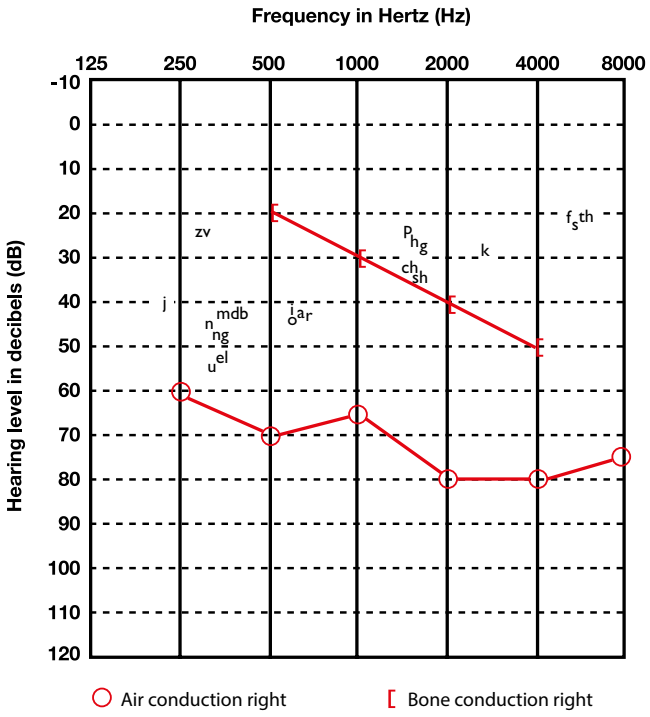
This audiogram shows a typical conductive deafness in a child's left ear. There are two lines – one shows the result of air-conduction tests (with headphones or earphones in the ears) marked by crosses, and the other shows bone-conduction results marked by square brackets ([). The bone-conduction test shows that the inner ear is receiving the signal clearly, but the air-conduction test shows that the sound is being blocked by fluid or another obstruction in the outer or middle ear. This child may have a temporary conductive deafness as a result of glue ear or a permanent conductive deafness.



Sensorineural deafness in the right ear

This audiogram shows a sensorineural deafness in the right ear. You can see that both the air- and bone-conduction tests give similar results.

Mixed deafness in the right ear



This last audiogram gives an example of mixed deafness in the right ear. Both the bone-conduction and air-conduction tests show that there is a hearing loss. Because the results are very different, this child has more than one cause of deafness.

6. Hearing tests with hearing aids

If your child does have a hearing loss, they may be fitted with hearing aids. Visual response audiometry, soundfield audiometry and speech testing can all be performed while wearing hearing aids, and the results will provide some information about what your child can hear with them. When these results are written down, they are called 'aided responses'. 'Real ear measurements' will also be used to make sure the hearing aid's settings are as close as possible to your child's hearing loss.

Real ear measurements (REM)

Your child's hearing aids will be programmed for their individual hearing loss. Two children with identical hearing losses and identical hearing aids will have slightly different prescriptions. This is because the size of each child's ear canal will be different, and this can alter the signal (or frequency response) coming from the hearing aid. The audiologist will use a probe tube microphone to take measurements in your child's ear canal to make sure that the hearing aid is set correctly.

This type of testing is not suitable for children who use bone conduction hearing aids.

Other ways to assess the benefit of hearing aids

Your audiologist or Teacher of the Deaf (ToD) will go through a questionnaire or checklist with you and your child to find out how well your child listens in different situations with their hearing aid (for example, how they are at identifying different sounds at home, working in groups at school, or using the phone). If your child is very young, your observations using the Early Support Monitoring Protocol for deaf babies and children may be used (you can download this from **www.ndcs.org.uk/education_resources.** The results of these can be used to fine-tune the settings of the hearing aids if necessary.

7. Causes of permanent deafness

There are many reasons why a child can be born deaf or become deaf early in life. It's not always possible to identify the reason, but you may be offered more tests to try to find out the cause of your child's deafness.

Causes before birth (pre-natal causes)

Around half the deaf children born in the UK every year are deaf because of a genetic (inherited) reason. Deafness can be passed down in families, even though there appears to be no family history of deafness. About 70% of these deaf children won't experience any other problems. For the other 30%, the gene involved may cause other disabilities or health problems.

Deafness can also be caused by complications during pregnancy. Infections such as rubella, cytomegalovirus (CMV), toxoplasmosis and herpes can cause a child to be born deaf.

Causes in early childhood (post-natal causes)

Being born prematurely, severe jaundice or a lack of oxygen at the time of birth can all increase the risk of a child being born deaf or becoming deaf early on. Premature babies are often more prone to infections and some medications used to treat them are known to be ototoxic (damaging to the ear). Infections during early childhood, such as meningitis, measles and mumps, can be responsible for a child becoming deaf.

Occasionally, a head injury or exposure to loud noise can damage the hearing system.

For more information about the tests used to find out the cause of deafness, read chapter 8, page 24.

8. Medical tests used to help diagnose the cause of permanent deafness

The process to find out why a child is deaf is sometimes called an 'aetiological investigation'.

The tests listed in this section can find the reason for a child's deafness in 40% to 50% of cases. For the other 50% to 60% of cases it's not possible to find out why a child is deaf. If it's not possible to find out the cause of your child's hearing loss, it may be helpful for you to know what didn't cause it.

Doctors may sometimes suggest tests on other parts of your child's body, like the kidneys or heart, to help find out the cause or rule out conditions that can be associated with deafness. Deafness can be part of a syndrome. Syndrome is a medical term meaning a collection of symptoms or signs that commonly appear together.

It's important to know about any associated medical conditions so you can think about appropriate treatment or ways of managing the deafness or condition. However, these conditions are quite rare in deaf children and very rare in the population as a whole.

As with many services provided by the NHS, for some tests you may be asked to give your written permission first.

What happens when you see the doctor?

The doctor will take details of your child's medical history. This will include questions about the pregnancy and the birth, including any medication that was taken during the pregnancy and the mother's health before, during, and after the birth.

The doctor will ask you about your child's immunisations (routine baby jabs). In toddlers and older children, the doctor will ask about your child's development (including speech, language and milestones such as when your child was sitting, walking etc.). The doctor may also ask about whether your child has:

- had meningitis, mumps, measles or other illnesses
- been exposed to loud noises
- taken any prescribed medication
- suffered any head injuries
- had any ear infections
- had sight problems
- had balance problems.

All of these factors are important when investigating the cause of deafness.

The doctor will also ask about the hearing of other family members, on both sides of the family.

Physical examination

The doctor will look at your child's head and face and may take some measurements. The doctor will also look at your child's neck, skin, nails, arms, legs, chest, abdomen (tummy), eyes, mouth, palate (roof of the mouth) and ears. They are looking for any minor differences or signs (for example, tiny holes in the skin known as 'pits'), that may help to diagnose the cause of the deafness. The doctor will assess your child's development in relation to the expected ages and developmental stages (see your baby's personal child health record – the red book – for further information on this).

The doctor may ask close family members to have a hearing test, known as pure tone audiometry.

Imaging

Imaging is a term covering different ways of looking at parts inside the body (such as bones or major organs) and how they're working.

The doctor uses an MRI (magnetic resonance imaging) scan or a CT (computerised tomography) scan to look at the structures of the ear and hearing nerve. Both types of scan are commonly used with children who have deafness.

An MRI scanner

An MRI scan will show soft tissues including the brain and hearing nerve. It will show if the hearing nerve has developed normally. MRI scans use magnets and radio waves to produce detailed pictures of the inside of the body. There are no known side effects associated with this type of scan. An MRI scan can be carried out on a child from birth, but you should be given a choice of when the scan is done. If it's important for your child to have the scan, the doctor will explain this to you.

A CT scan will show the bony parts of the ear including the 'ossicles' (the three tiny bones in the middle ear) and the 'cochlea' (the inner ear). A CT scan will show if the bony parts have developed normally. The scan involves exposing your child to radiation in the form of X-rays. The level of radiation used is kept as low as possible to prevent damage to body cells, and the amount of radiation your child is exposed to depends on the number of images taken. It's generally accepted that there's little risk to health from one scan, but with repeated tests there's a risk that the radiation may damage body cells. The earlier in life your child is exposed to radiation, the greater the risk. You and the doctor may prefer to wait until your child is a little older. Your child's doctor will discuss this with you and answer any questions you may have.

Your child will need to be absolutely still for the time it takes to do a CT or MRI scan. Very young babies may be able to have these scans while they are asleep. Each hospital will have its own procedure, and your doctor will explain this to you. Children aged over three months will normally be given something to help them sleep. This may be a light sedative or a short general anaesthetic.

For children aged over two, you may be offered a stronger sedative or a general anaesthetic. Although modern anaesthetics are very safe, there are small risks associated with having an anaesthetic and your doctor should explain these to you. The risks of having an anaesthetic reduce as the child gets older. Usually, children over the age of five can lie still for the scan without needing a sedative or anaesthetic.

A further type of scan that may sometimes be used is a renal ultrasound. This is a scan that uses sound waves to create images of the kidneys. It's similar to the scans used during pregnancy. It's only likely to be used to rule out a rare syndrome or if there's a family history of kidney problems. There are no risks associated with this type of scan.

Your doctor may advise that one or more of the above scans are done at an early stage, for example, if:

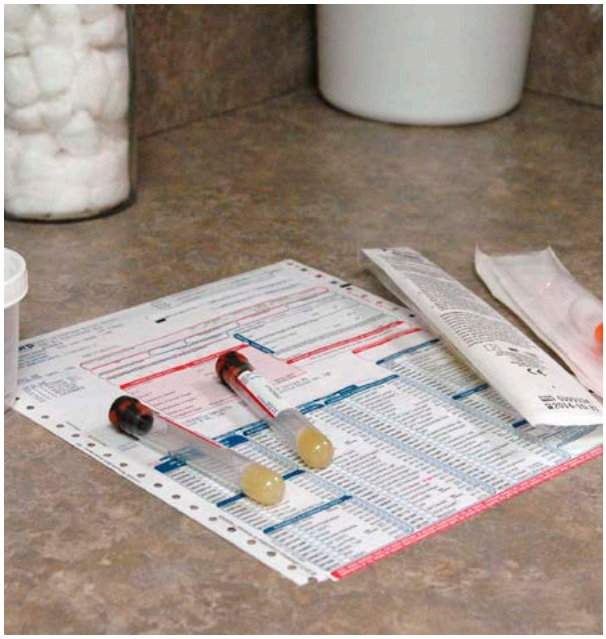
- your child has had meningitis
- the deafness is getting worse or changing over time
- there are characteristic features that may suggest your child's deafness could be part of a syndrome.

In these examples, your doctor will need to look closely at the structure of the cochlea (inner ear), balance organs and hearing nerve to be able to give you advice on possible management or treatment options.

Electrocardiography (ECG)

An ECG is a recording of the rhythm and electrical activity of the heart. There's a very rare syndrome linking severe and profound hearing loss to a heart problem. If this heart problem is found, it can be treated.

Depending on your child's and your family's medical history, your doctor may advise an ECG for a small number of children with severe to profound deafness to help rule out this condition. There are no risks associated with having this test.



Blood and urine tests

Depending on your child's and your family's medical history, the doctor may ask for one or more routine blood and urine tests. These tests can help doctors to identify the cause of the deafness.

An example of a blood or urine test your child may be offered is an infection screen. An infection screen is used to look for certain infections that sometimes result in deafness. Some of these tests will give useful results only if they're carried out in the weeks or months shortly after your child's birth. When the child is older, an infection screen may give a negative result and rule out an infection being the cause of the deafness. However, a positive result later in life may not give useful information as your doctor couldn't prove that the infection was there at birth and so caused the deafness.

There are several infections that can cause deafness in babies if the mother contracts them when pregnant. The most common infection causing babies to be born with deafness is CMV (cytomegalovirus). CMV is very common in the general population and does not normally cause any illness. However, it can affect the baby if the mother catches it while pregnant. CMV infection in an unborn baby is called congenital CMV (cCMV). 'Congenital' means from birth. About one baby in every 150 is born with CMV and some of these babies can be affected by it. cCMV can cause deafness or, very occasionally, it can affect a child's development. It causes about 10% to 20% of deafness in children in the UK. Doctors can test for CMV in the first few weeks after birth, or, for older children, look for signs of the CMV infection in the heel-prick blood test cards which are stored after birth. Deafness following infection can sometimes get worse over time.

If a young baby is found to have the CMV infection, it may be possible to give treatment that may prevent the deafness from increasing. This must be given in the first four weeks of being born.

For more information read our factsheet *Congenital Cytomegalovirus and Deafness*.

Less common infections that can cause deafness include rubella (German measles), toxoplasmosis and syphilis. Your doctor will be able to give you more information about these infections. All the blood tests are usually done using one blood sample.

Ophthalmology (eye test)

All children learn from what they see and hear around them. Children who are deaf rely on their eyesight even more than other children do. Up to 40% of children with sensorineural deafness also have an eye problem. This may simply mean the child will need to wear glasses when they are older, but an eye test can also help to diagnose a syndrome associated with deafness. As babies can't tell us what they can see, a developmental assessment of the eye is done to make sure the eyes are healthy. Sometimes special eye tests are used. Your opthamologist (eye doctor) will discuss this with you if necessary. It's recommended that all children diagnosed with deafness are referred for an eye test and have regular eye tests throughout their childhood.

For more information about eye tests visit **www.ndcs.org.uk/visioncare**

Genetic counselling

Just as children inherit features such as hair colour or eye colour from their parents, sometimes deafness is inherited.

If your child's deafness could have a genetic cause, you should be given the chance to discuss this with a trained genetics counsellor. Genetic counselling gives families information about:

- the cause of a range of inherited conditions
- how an inherited condition might affect the child and family in the future
- how likely you are to have another child with the same condition.

Some families find it helpful to know whether the deafness and any other medical condition were inherited. Other families prefer to wait until their children are grown up and able to decide for themselves.

For more information read our resource *Genetic Counselling – Information for families*.

Genetic testing

You may be offered a genetic test. This will involve your child, and possibly other family members, having a blood test.

The blood sample will be used to look for the gene or genes known to be linked with deafness, for example Connexin 26. Not all the genes related to deafness have been identified and for most there is not yet a routine test. This means that even if the deafness is inherited, it may not be possible to confirm this with a genetic test at the moment. About 50% of cases of permanent deafness in children have a genetic cause. In about 30% of children with a genetic deafness, the deafness is part of a syndrome. Again, some of these syndromes can be confirmed with a genetic test, but many can't.

Some children have a rare genetic deafness that can get worse if they are given certain medications. It's possible to have a genetic test to identify whether they have this gene. If they do, then this information will help inform you and your doctor if, in the future, your child needs treatment. Your doctor may advise that this test is done at an early stage depending on your child's history.

Your doctor may refer you to a clinical geneticist or a genetic counsellor. Alternatively, they may discuss genetic testing with you themselves. Individual health authorities decide whether to offer routine genetic testing through the audiology or ENT department. Some areas offer genetic testing through this route only if it would provide a direct benefit to the child being tested. For example, if the child has an inherited medical condition it may be of direct benefit to have a genetic test. This is because the results may help to identify treatments that should be made available to the child. Your doctor will discuss the options available in your area with you. If you want to ask more about genetic testing, you can ask to be referred to your local clinical genetics service and your doctor will discuss with you if this is appropriate.

For more information on the genetics of deafness read our resource *Genetic Counselling – Information for families*.

What next?

Some or all of the tests in this section will be offered to your child, not necessarily in the order given here. Your doctor will give you more information which will help you decide whether you want your child to have the tests, and the best time for these to be carried out. Test results that may offer immediate benefit to a baby are best done at an early stage, for example when the infection screen is carried out.

Some tests (for example, some routine genetic testing) will not offer an immediate or direct benefit to a baby and may be delayed. Your doctor will tell you if particular tests should be done (for example, if they think that your child's deafness is part of a syndrome). You and your doctor can use this information when planning how to manage the hearing loss and any associated condition in the future.

Most of the tests are not urgent. It's important that you feel comfortable with your child having them. If you have any worries, you can talk about them with your doctor.

9. Finding medical information online

The internet can be a very useful source of information for families, but the number and types of websites can be confusing.

This chapter will help you decide whether a website provides relevant, high quality information.

What to look for on support group websites

Support groups' websites are a way of getting in touch with other people. Before you start using a website, have a think about the points below.

- Can you find clear contact details for the organisation that set up the website?
- If there are email lists, bulletin boards and chat rooms, you're likely to be in touch with people who are genuine. But remember, some may have extreme views.
- Check that your personal information will be kept secure and not shared with others.
- See if your personal details are being recorded.
- Can you contact the website manager to report technical problems and give feedback about the site?

How reliable is the medical information?

When you're looking at medical information online, have a think about the following.

- Check the author's name, job title, workplace, and any formal or professional qualifications.
- Has the information been certified as reliable health information by The Information Standard? (It should carry its quality mark – see the back cover of this resource.)
- Check the date the information was put on the site. Medical information can become out of date very quickly.
- Is the information aimed at getting you to buy something?
- Does the information acknowledge that specific conditions affect people in different ways, ranging from mild to severe?
- Check whether the information is based on a person's own experiences. How the condition affects your child may differ from other people's experiences.
- Does the information sound extreme?

Who is the website for?

Websites are aimed at different groups of people (for example, professionals, academics, members of the public). Detailed academic research can be confusing, and may not be helpful. Think about who the website is aimed at and how useful the information will be.

Who produced the website?

Websites can be set up by anybody, from respected organisations and experts to people with extreme views and companies trying to sell you something.

Have a think about the following.

- Look for the name, address and contact details of the organisation.
- Does the organisation have a registered charity number (if relevant)?
- What are the aims and purpose of the organisation?
- Check the names and qualifications of any professionals contributing to the website. Is there an advisory panel or review group?
- Websites should state clearly if the information is based on people's personal experiences.
- Websites sponsored by commercial organisations may be biased towards certain treatments or products. Adverts that appear on a website might also reflect this.

Websites from outside the UK can be useful, but may refer to medicines using different names from those used here, or ones not licensed for use in this country. There may be different medical practice and treatments in other countries.

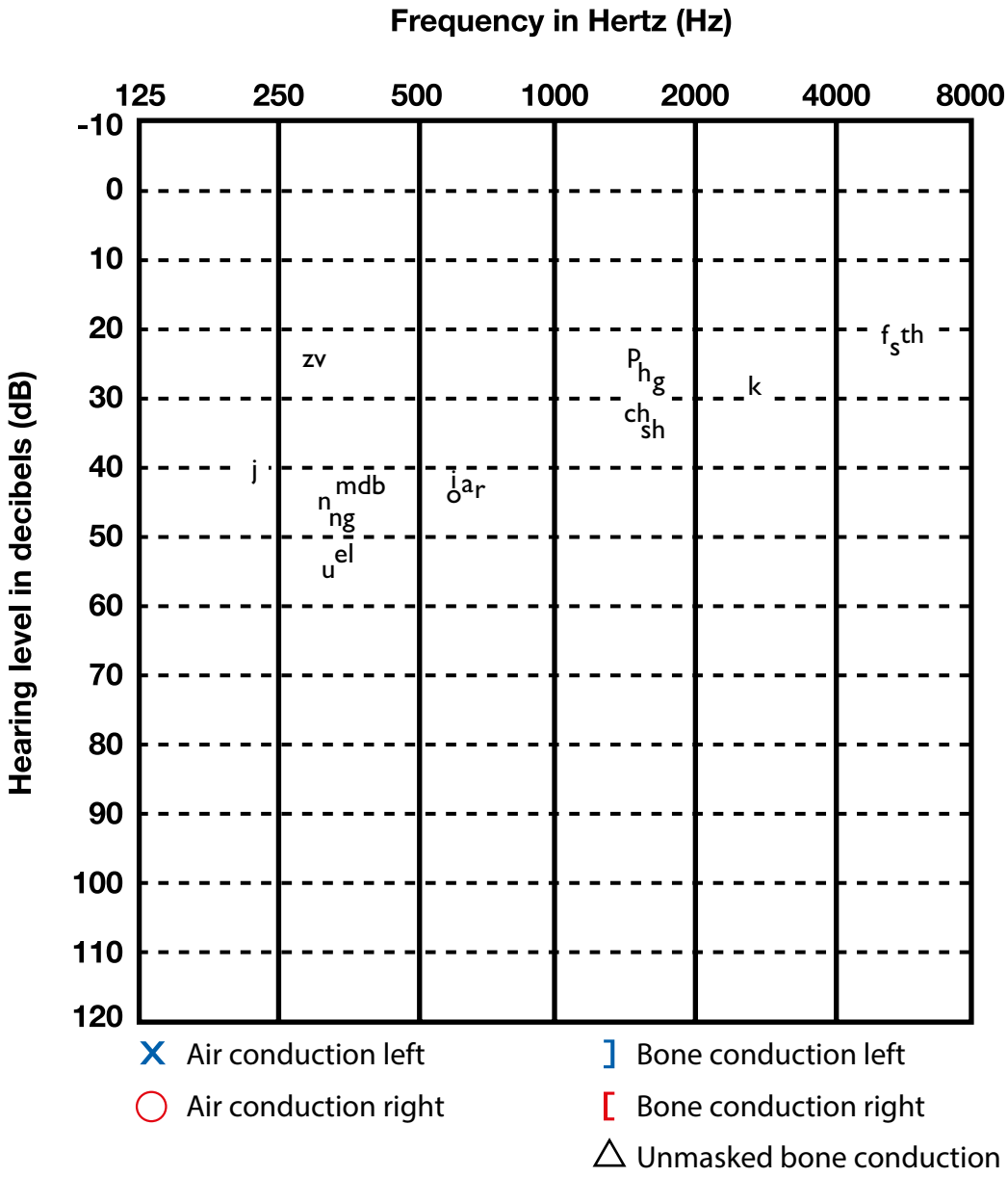
10. Audiology appointments questions list

To help you understand the hearing tests that are being carried out on your child and what the results mean, you may want to ask some of the questions below at your child's audiology or ear, nose and throat (ENT) appointment.

1. Could I have a copy of the most recent hearing test and the report?
2. What tests have been carried out? Will there be a need for further testing?
3. What type of hearing loss does my child have (sensorineural, conductive, or mixed)? Please explain what this means.
4. Will my child need to be referred to another health professional? For example, an eye doctor or a geneticist?
5. Do you think our family should have genetic counselling?
6. Does my child need other tests? E.g. scans (CT, MRI), heart (ECG), blood, urine tests or eye tests. What will these tests tell you about my child's hearing loss?
7. Will my child's hearing loss improve or progress (get worse)?
8. What caused my child's hearing loss?
9. Do we and other members of our family need to have our hearing tested?
10. What options are there for my child? (e.g. grommets, hearing aids or implants).
11. Will a hearing aid help my child to hear?
12. When will my child be fitted with hearing aids?
13. Is a cochlear implant an option for my child? Where can I get more information?
14. Will the hearing loss affect my child's development?
15. Will my child be able to hear me?
16. How often will my child come back for a follow-up appointment?

11. Audiogram for your child's test results

Ask your audiologist to print off a copy of your child's hearing test results for you or plot their results on this chart. This can help you explain what your child can and cannot hear, with and without their hearing aids.



The National Deaf Children's Society is the leading charity dedicated to creating a world without barriers for deaf children and young people.

Freephone Helpline: **0808 800 8880** (voice and text)

helpline@ndcs.org.uk

www.ndcs.org.uk/livechat

www.ndcs.org.uk

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Full references for this resource are available by emailing **informationteam@ndcs.org.uk**
Give us your feedback by emailing your comments to **informationteam@ndcs.org.uk**

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